HEAD AND NECK VASCULAR ANOMALIES

A Practical Case-Based Approach

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Preface

A better understanding of the nature and source of vascular anomalies has vitalized an interest in this field among numerous disciplines. The language used to describe these lesions is now coherent across specialties and allows for treatment algorithms to be unified. However, each vascular tumor and malformation has a unique management profile based on its type, size, and location as well as disciplines involved. Head and neck vascular anomalies are no exception to this rule and are the subsequent motivation behind this text.

We designed *Head and Neck Vascular Anomalies: A Practical Case-Based Approach* with the goal to provide hands-on, step-by-step, management algorithms for specific vascular anomalies of the head and neck encountered in daily practice. This is a condensed, multidisciplinary, practical guide for both simple and complex lesions. Our colleagues in otolaryngology, dermatology, pediatric surgery, plastic surgery, oncology, and interventional radiology have all contributed amazing cases with clinical detail, scientific evidence, and therapeutic options.

In each chapter, the initial steps to diagnose a vascular lesion are followed by a recommended treatment in a case-based format with photographs, radiographic imaging, and alternative therapies. All cases are based upon current literature with the aim to give state-of-the-art information on the majority of head and neck vascular anomalies. Medical, radiographic, and surgical techniques for frequently encountered and more difficult vascular anomalies are described.

This text is designed to be a reference guide. As you will see, each case follows a consistent and relatively rigid presentation outline. This style is meant to provide clarity, brevity, and simplicity to the reader. As a result, redundancies may be encountered for similar anomalies. For this we apologize, but frankly, we did not design the text to be read from cover to cover. Actually, we hope the reader can simply turn to a chapter and capture a complement of knowledge required to help their specific patient.

Of note, we also did not filter out any author or discipline bias in the chapters. In essence, the authors were allowed to express their opinion and therapeutic approach to their assigned case with the requirement to provide treatment alternatives. This decision was made to maintain the authenticity of opinion that is frequently found in the multidisciplinary field of vascular anomalies.

We, thereby, humbly submit to you *Head and Neck Vascular Anomalies: A Practical Case-Based Approach.* With an increasing number of vascular anomaly centers, patients, and interest in the field, we hope you find this text important to your everyday practice and a valuable aid for your patients.

—Gresham T. Richter and James Y. Suen

Introduction to Vascular Anomalies

Lauren A. Kilpatrick

HISTORY

Vascular anomalies are complex disruptions in normal vascular development that may affect as many as 1 in 10 people. Although they may occur anywhere on the body, the head, neck, and face are common locations for their appearance. Frequently referred to as vascular birthmarks, vascular anomalies are considered benign vascular lesions that vary drastically in their presentation, growth, and treatment options. Because they are encountered by many medical and surgical disciplines, the nomenclature for these lesions has been historically disjointed and confusing. Multiple terms referred to the same anomaly, while in contrast, the same term may have been used for fundamentally different anomalies. This frequently led to inadequate or inappropriate treatment algorithms. Fortunately the Society for the Study of Vascular Anomalies (ISSVA) was established in 1992 after 16 years of biennial international workshops. This multidisciplinary organization dedicated themselves to understanding vascular anomalies and to developing a consensus on classifying and improving the clinical care of patients with these disorders.

Prior to 1982, classification for benign vascular lesions was largely based on pathology, similar to the histogenetic system used for soft tissue tumors.[1] Vascular lesions were divided into localized or diffuse forms and described based on their size, predominant vessel, and type of tissue involved.[2] In 1982, Mulliken and Glowacki proposed a reorganization for the nomenclature of benign vascular lesions with the basic premise of differentiating vascular tumors from malformations.[3] Vascular tumors were characterized by rapid growth and sometimes slow involution, attributed histologically to hyperplasia during proliferation and fibrosis during involution. Vascular malformations were stated to be present at birth and thought to grow proportionally with the patient, demonstrating an error of vascular morphogenesis but with a normal rate of cell turnover and mitosis.[3–5]

CLASSIFICATION

The ISSVA classification system was adopted in 1996 as the primary clinical tool in diagnosing vascular anomalies and based on the system published by Mulliken and Glowacki.[3] Proper history and physical examination can accurately diagnosis a vascular lesion in 96% of patients using the ISSVA classification.[6] However, the World Health Organization (WHO) continues to distinguish between vascular lesions involving the skin versus soft tissue and this classification is used primarily by pathologists.[7–8] The ISSVA classification essentially divides vascular anomalies into two broad categories based upon clinical history and histology: vascular tumors and vascular malformations. Vascular malformations are then subdivided into slow-flow, fast-flow, or complex-combined lesions (Figure 0–1). Various types of vascular tumors are also clarified of which infantile hemangiomas are the most common. The 2014 updated ISSVA classification is summarized in Table 0–1.[5]

DIAGNOSIS

Clinical history and examination are key to proper diagnosis of vascular anomalies and are highly predictive.[6] An algorithm of simple questions can help target the diagnosis at first presentation (Figure 0–2). Malformations are most commonly present at birth while infantile hemangiomas are either absent or at

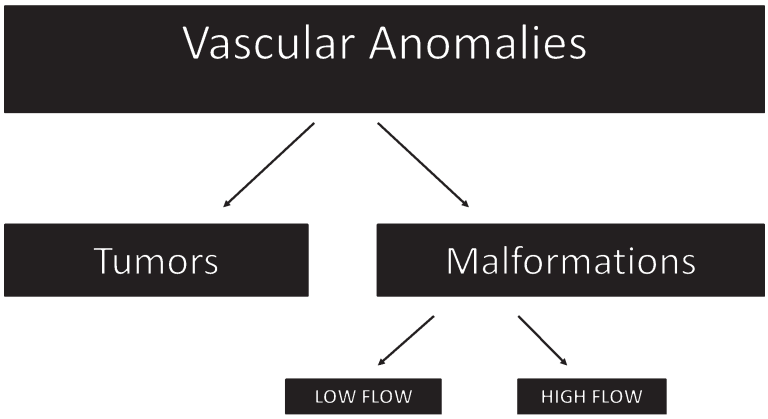

Figure 0–1. Simple schematic of the ISSVA classification of vascular anomalies.

Table 0–1. Abbreviated ISSVA Classification System of Vascular Anomalies

Vascular tumors	Infantile hemangioma	
	Congenital hemangioma	Noninvoluting (NICH) Rapidly involuting (RICH)
	Other tumors	Tufted angioma Kaposiform hemangioendothelioma Spindle cell hemangioendothelioma Other hemangioendotheliomas
	Dermatologic acquired tumors	Pyogenic granuloma Glomeruloid hemangioma Other acquired tumors
Vascular malformations	Slow flow	Capillary malformation (CM) Venous malformation (VM) Lymphatic malformation (LM)
	Fast flow	Arterial malformation (AM) Arteriovenous fistula (AVF) Arteriovenous malformation (AVM)
	Complex-combined	CVM CLM LVM CLVM AVM-LM CM-AVM

Note. Adapted from the International Society for the Study of Vascular Anomalies. Retrieved from https://issva.clubexpress.com/docs.ashx?id=178348

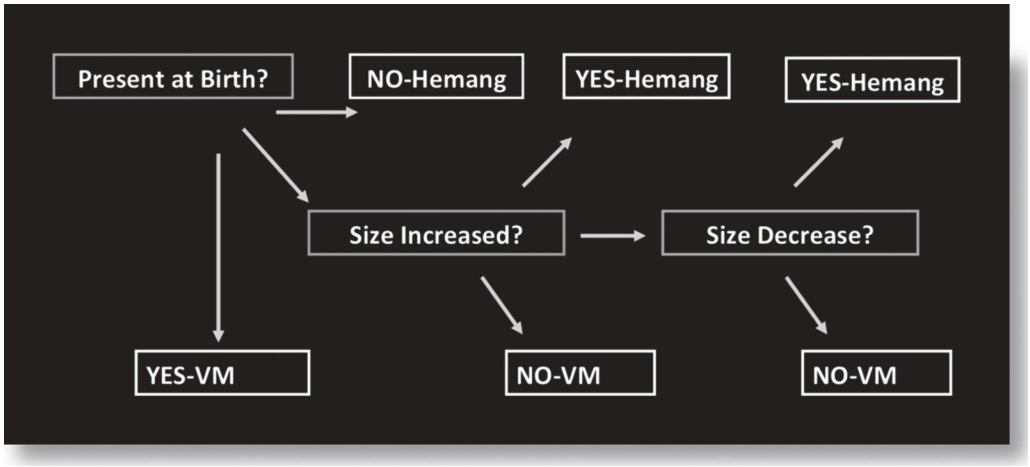

Figure 0–2. Algorithm of questions to help differentiate a vascular tumor from a vascular malformation by patient history.

their smallest size at birth. Other vascular tumors, such as congenital hemangiomas and hemangioendotheliomas are present at birth but have a distinctive growth pattern compared to malformations. Proliferation and eventual involution of the lesion also indicate the presence of an infantile hemangioma, whereas vascular malformations characteristically grow proportionate to the patient with limited changes in size during childhood except during episodes of trauma, infection, and hormonal changes. Gradual expansion of vascular malformations is the general rule but will lead to significant aesthetic and functional issues over time. Complex or large vascular malformations can also have associated coagulopathies, vascular steal, and hypertrophic disorders with life-threatening issues.

On physical examination, vascular tumors more commonly have well-defined borders though some can be infiltrative. Malformations can be focal or multifocal (diffuse). The multifocal malformations will have poorly defined margins that contribute to a higher rate of recurrence following treatment. Malformations may be compressible with temperature variation based on their contributing vessel type (eg, arteriovenous malformations are typically warm and pulsatile).[9]

Imaging studies are often beneficial in the evaluation of a vascular anomaly. Specifically, ultrasound and magnetic resonance imaging (MRI) are the most useful radiographic tools used in the diagnosis of vascular anomalies. Ultrasound is advantageous as it is noninvasive, inexpensive, easily accessible, and does not require sedation. The limitation of ultrasound is its ability to evaluate deep tissues. MRI with gadolinium contrast has excellent soft tissue resolution but is costly and may require sedation, particularly in pediatric patients. Both modalities have no radiation exposure. Table 0–2 lists typical features of vascular anomalies on ultrasound, and Table 0–3 lists these features on MRI. Computed tomography (CT) is limited in its ability to delineate soft tissue densities and requires radiation, though it can be advantageous for evaluating bone involvement. CT arteriography is also useful to delineate an arteriovenous malformation and can be performed much quicker than an MRI. Angiography (± venography) is the ideal study in the evaluation of arteriovenous malformations and may provide access for intervention.[10–12] An arteriogram is not indicated or useful with venous malformations.

Histologic differences also exist between vascular tumors and malformations. North et al. discovered that glucose transporter protein, GLUT1, is expressed by endothelial cells of infantile hemangiomas but is not seen in vascular malformations.[13] GLUT1 is found in placentas and no other tissue in the body, which suggests that infantile hemangiomas may be metastases from placenta. Con-

genital hemangiomas are also negative for GLUT1 expression. Markers of cellular proliferation, most notably vascular endothelial growth factor (VEGF), are increased in proliferating hemangiomas while vascular malformations rarely express high levels of VEGF.[14] Although histologic evaluation may not be necessary for all cases, diagnosis for atypical presentations of vascular lesions may be improved with pathologic findings. D2-40 is a good marker for lymphatic endothelium to help diagnosis a lymphatic malformation. Arteriovenous malformations can be differentiated from other vascular malformations by their expression of CD105.[15] A list of genetic mutations associated with their vascular malformation phenotype can be found in Table 0–4.

Table 0–2. Ultrasound (US) Characteristics of Vascular Anomalies

	Standard US	Doppler US
Hemangioma	Solid Homogeneous Well circumscribed	Hypervascular Arterial and venous waveforms
Slow-flow malformation	Compressible Echogenic Phleboliths (VM) Fluid levels/cystic spaces (LM)	Monophasic waveform No flow
High-flow malformation	Vascular cluster Poorly demarcated	Arterial waveforms Loss of normal venous dampening

Table 0–3. Magnetic Resonance Imaging (MRI) Characteristics of Vascular Anomalies

	T1	T2	Postcontrast Enhancement
Hemangioma	Isointense	Hyperintense	+
Slow-flow malformation			
• Venous malformation	Isointense	Hyperintense	+ (patchy)
• Lymphatic malformation	Hypo- to isointense	Hyperintense	− (may be + in septae)
High-flow malformation	Flow voids	Flow voids	−

Table 0–4. Genetic Mutations Specifically Identified in Various Vascular Malformations

Phenotype	Identified Mutations
Capillary malformation (PWS)	GNAQ
Venous malformation	TIE2
Capillary-arteriovenous malformation	RASA1

REFERENCES

1. Enzinger FM, Weiss SW. *Soft Tissue Pathology*. St Louis, MO: CV Mosby; 1983:1–12.
2. Enzinger FM, Weiss SW. *Soft Tissue Pathology*. St Louis, MO: CV Mosby, 1983:379–421.
3. Mulliken JB, Glowacki J. Hemangiomas and vascular malformations in infants and children: A classification based on endothelial characteristics. *Plast Reconstr Surg*. 1982;69:412–422.
4. Kransdorf MJ, Murphey MD, Fanburg-Smith JC. Classification of benign vascular lesions: History, current nomenclature, and suggestions for imagers. *Amer J Roentgenol*. 2011; 197:8–11.
5. Enjolras O, Wassef M, Chapot R. Introduction: ISSVA classification. In: *Color Atlas of Vascular Tumors and Vascular Malformations*. Cambridge, UK: Cambridge University Press; 2007:1–12.
6. Finn MC, Glowacki J, Mulliken JB. Congenital vascular lesions: Clinical application of a new classification. *J Pediatr Surg*. 1983; 18:894–900.
7. Calonje E. Haemangiomas. In: Fletcher DM, Unni KK, Mertens F, eds. *WHO Classification of Tumors, Pathology, and Genetics: Tumors of Soft Tissue and Bone*. Lyon, France: IARC Press, 2002:156–158.
8. Sangueza OP, Kasper RC, LeBoit P, et al. Vascular tumors. In: LeBoit PE, Burg G, Weedon D, Sarasin A, eds. *WHO Classification of Tumors, Pathology, and Genetics: Skin Tumors*. Lyon, France: IARC Press, 2006:233–246.
9. Waner M, Suen JY. Management of congenital vascular lesions of the head and neck. *Oncology*. 1995;9:989–994,997.
10. McCafferty IJ, Jones RG. Imaging and management of vascular malformations. *Clin Radiol*. 2011; 66:1208–1218.
11. Behr GG, Johnson C. Vascular anomalies: Hemangiomas and beyond—Part 1, fast-flow lesions. *AJR Am J Roentgenol*. 2013; 200:414–422.
12. Behr GG, Johnson C. Vascular anomalies: Hemangiomas and beyond—Part 2, slow-flow lesions. *AJR Am J Roentgenol*. 2013; 200:423–436.
13. North PE, Waner M, Mizeracki A, Mihm MC Jr. GLUT1: A newly discovered immunohistochemical marker for juvenile hemangiomas. *Hum Pathol*. 2000; 31:11–22.
14. Takahashi K, Mulliken JB, Kozakewich JPW, Rogers RA, Folkman J, Ezekowitz RA. Cellular markers that distinguish the phases of hemangiomas during infancy and childhood. *J Clin Invest*. 1994; 93:2357–2364.
15. Hou F, Dai Y, Dornhoffer JR, Suen JY, Fan CY, Saad AG, Buckmiller LM, Richter GT. Expression of endoglin (CD105) and endothelial nitric oxide synthase in head and neck arteriovenous malformations. *JAMA Otolaryngol Head Neck Surg*. 2013; 139(3):237–243.

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—Gresham T. Richter

My thanks to Gresham Richter, MD, for his concept for this book which is unique and practical and hopefully will be a great resource for Medical Practitioners and patients and their families who suffer from this poorly understood medical problem.

—James Y. Suen

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To all patients afflicted by a vascular anomaly or birthmark in a world that poorly understands. To their families who also live with the anomaly every day and support their loved one's self-concept and often complex treatment plans. Also, we dedicate this book to the early leaders of the field and a new generation of physicians and scientists aiming to find a cure.

To my sweet wife Anna, and our three wonderful children August, Charlie, and Lucy whom I love dearly. To my mother for teaching me how to write and Jeanne and Ladd Goesl for setting such a good example.

—Gresham T. Richter

To my family, Karen, Brent, Tiffany, Bradley and Jessica, Brennan, and my two beautiful granddaughters, Sophia and Vivian. Thanks for understanding my passion for helping my patients.

—James Y. Suen

1

Infantile Hemangiomas

BASIC TENANTS AND INTERVENTIONS

Gresham T. Richter

Basic Tenants

Infantile hemangiomas (IHs) are the most common vascular tumor. They are composed of proliferating immature endothelial cells that express histologic marks found on placental blood vessels (GLUT-1, Lewis Y Antige, FcyRII, and merosin).[1] IHs are thought to be sporadic events although family lineage has been reported.[2] Coincidentally, IHs are also the most common tumor of infancy and are present in approximately 5% of the population.[3] They have a higher prevalence in females, Caucasians, and premature, and low birth weight infants.[4] They also occur more frequently in infants from mothers with early trimester bleeding, preeclampsia, and placental anomalies.

Infantile hemangiomas are rarely present at birth but early blanching or macular erythema of the skin may be a precursor to their later development. They may present anywhere on the body but involve the head and neck in over 60% of cases. Eighty percent of IH grow within the first 3 months of life and continue to grow up to 1 year of age.[5] IHs undergo predictable proliferative, quiescent, and involution

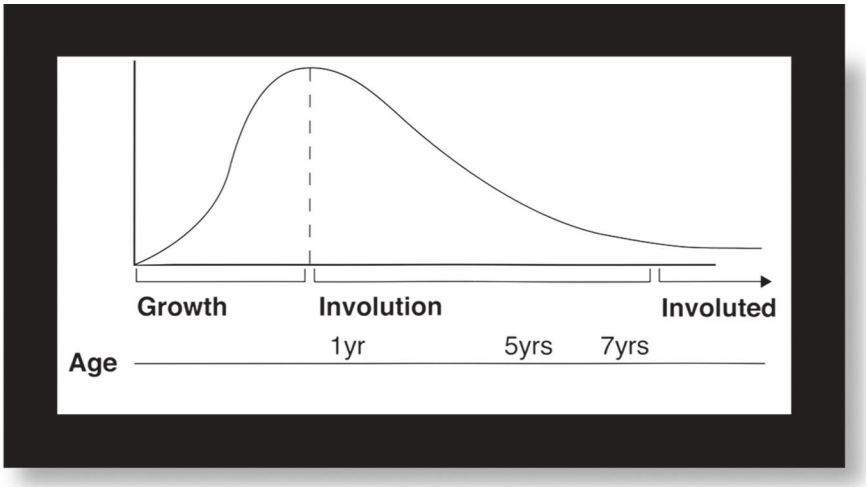

Figure 1–1. Typical growth phases of infantile hemangiomas.

phases as seen in Figure 1–1. The majority of IHs are thought to involute completely by 7 years of age. This natural history can help the clinician differentiate IHs from other congenital lesions and guide management decisions.

The classification of IHs is rather complex. They are first determined to be either focal or segmental. Focal IHs have discrete borders and further characterized as either superficial, deep, or compound. Early nomenclature has been supplanted by this new terminology to describe the majority of IHs (Table 1–1). Superficial and compound hemangiomas present with dark red cutaneous staining in a cobblestone pattern. Compound hemangiomas contain a subcutaneous component whereas deep hemangiomas do not involve the skin and present as a protuberance with an overlying blue skin discoloration (Figure 1–2). It is extremely rare for a focal IH to involve muscle or penetrate beyond subcutaneous fat. An exception is a parotid IH, the most common nonepitheliod tumor of the gland, which is frequently deep.

Problematic focal IHs typically involve the lip, eyelid, orbit, and subglottis where aesthetic and functional problems occur during the rapid proliferative phase. Sixteen percent of infants with 5 or more focal IHs will also have hepatic involvement and should undergo abdominal ultrasound.[6] Segmental IHs have a more complex growth pattern that their focal counterpart. In the head and neck, segmental IHs follow a trigeminal nerve (V) distribution. They are diffuse, compound, and maintain irregular borders. More than one facial subunit is frequently involved. They usually penetrate into deep fascial planes of the head and neck. The beard distribution IH (V3) is most commonly described.[7] These involve the lower lip, chin, neck, and preauricular areas and are frequently accompanied with ulceration. Sixty-three percent of segmental beard distribution will involve the subglottis and require airway endoscopy. All patients with segmentally distributed IHs should undergo systematic evaluation for PHACES (posterior fossa malformations, hemangiomas, arterial lesions, cardiac abnormalities, eye abnormalities, sternal cleft) syndrome.

The cause of IHs remains unclear but is postulated to either be ectopic placental tissue or an endo-

Table 1–1. Old and New Nomenclature for Infantile Hemangiomas

Old Nomenclature	New Nomenclature
Strawberry or Capillary Hemangioma	Superficial Hemangioma
Cavernous Hemangioma	Deep Hemangioma
Capillary Cavernous Hemangioma	Compound (Mixed) Hemangioma

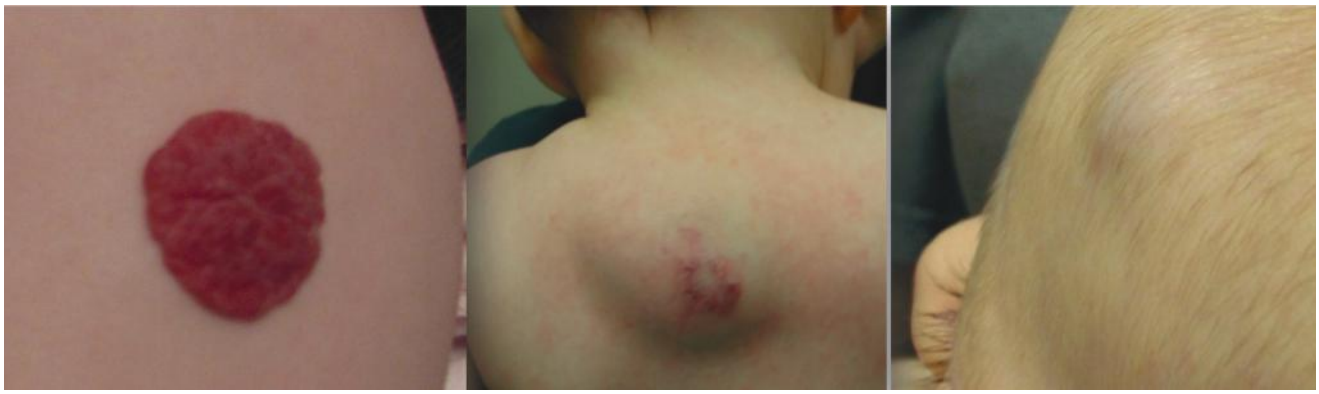

Figure 1–2. Focal hemangiomas described as superficial, compound, or deep (*left to right*).

thelial progenitor stem cell.[8] IHs are not associated with increased morbidity or mortality except in the very large hemangiomas that may rarely cause high output heart failure.

Intervention

Because of their natural involution, IHs were historically managed with observation alone. Although many resolve spontaneously others will cause significant functional and disfiguring consequences. Problematic hemangiomas are defined as those leading to significant events affecting the future life of the child. Most problematic events from IHs occur during the proliferative phase and include ulceration, bleeding, pain, vision disturbance, airway compromise, and feeding difficulties. However, late and deforming sequelae also occur to include scarring, telangiectasias, and fibrofatty residuum (Figure 1–3). Many cease to improve after 4 years of age and up to 69% of IHs will leave residual lesions.[9] At least 10% of IHs persist beyond 9 years of age. The age of self-recognition occurs around 4 years of age and must be considered in the treatment hemangiomas during their early phase of growth. Although it is difficult to predict future consequences for each lesion, early observation for rapid growth, protuberance, segmental disease, and functional compromise will help guide appropriate therapy.

Both surgical and medical interventions are available in the treatment of IHs. These include surgical excision, laser therapy, topical therapy, intralesional corticosteroids, systemic corticosteroids, systemic beta-blockers, and vincristine chemotherapy. Each of these therapeutic modalities is discussed in the following case presentations. Every IH has a unique profile that governs its treatment and is typically based on location and risk of aesthetic and functional compromise. Management during the proliferative phase generally will lead to the best final outcome. However, many IHs require multimodal therapy of which the final treatment occurs during the involution period. Absolute indications for early intervention include an impact on vital structures, active or impending functional impairment, the possibility of permanent scarring, large segmental facial hemangiomas, and ulcerative lesions.

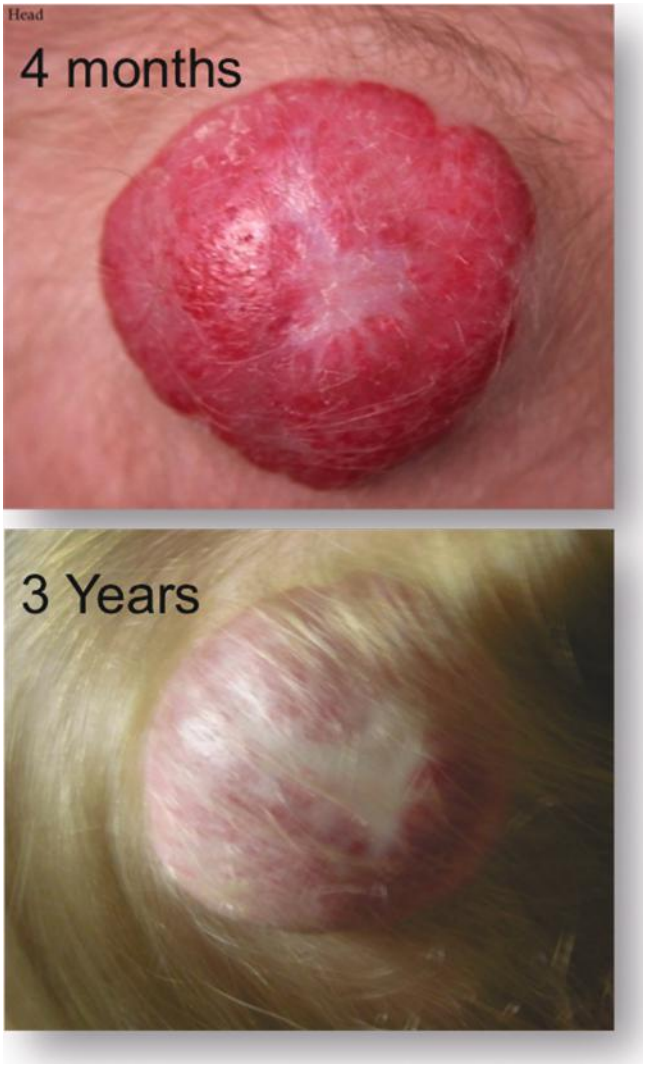

Figure 1–3. Focal scalp hemangioma at 4 months and seen again, untreated, at 3.5 years with resultant residuum that will require intervention.

References

1. North PE, Waner M, Mizeracki A, Mihm MC, Jr. GLUT1: a newly discovered immunohistochemical marker for juvenile hemangiomas. *Hum Pathol.* 2000;31:11–22.
2. Blei F, Walter J, Orlow SJ, Marchuk DA. Familial segregation of hemangiomas and vascular malformations as an autosomal dominant trait. *Arch Dermatol.* 1998;134: 718–722.
3. Dickison P, Christou E, Wargon O. A prospective study of infantile hemangiomas with a focus on incidence and risk factors. *Pediatr Dermatol.* 2011;28:663–669.

4. Haggstrom AN, Drolet BA, Baselga E, et al. Prospective study of infantile hemangiomas: demographic, prenatal, and perinatal characteristics. *J Pediatr.* 2007;150:291–294.
5. Chang LC, Haggstrom AN, Drolet BA, et al. Growth characteristics of infantile hemangiomas: implications for management. *Pediatrics.* 2008;122:360–367.
6. Horii KA, Drolet BA, Frieden IJ, et al. Prospective study of the frequency of hepatic hemangiomas in infants with multiple cutaneous infantile hemangiomas. *Pediatr Dermatol.* 2011;28:245–253.
7. Orlow SJ, Isakoff MS, Blei F. Increased risk of symptomatic hemangiomas of the airway in association with cutaneous hemangiomas in a "beard" distribution. *J Pediatr.* 1997;131:643–646.
8. Yu Y, Flint AF, Mulliken JB, Wu JK, Bischoff J. Endothelial progenitor cells in infantile hemangioma. *Blood*. 2004; 103:1373–1375.
9. Luu M, Frieden IJ. Haemangioma: clinical course, complications and management. *Br J Dermatol*. 2013;169:20–30.

CASE STUDY 1–1. ANTERIOR NECK HEMANGIOMA

Abby R. Nolder

Representative Case

A 2-month-old, former 26-week preterm newborn male was referred to the pediatric otolaryngology clinic for evaluation of middle ear pathology following a failed newborn hearing screen. During that visit, the patient's mother expressed concern about a growing mass under the child's chin. It was not present at birth but had been rapidly progressing over the last several weeks. He had a history of intubation for 2 days in the neonatal intensive care unit but had no associated airway symptoms. He was having some feeding difficulties that seemed to be worsening as the mass increased in size.

On physical examination, he was found to have a 4-cm, soft, mobile, cystic appearing submental neck mass with faint blue discoloration of the overlying skin (Figure 1–4). He also had a 3 × 4-cm compound, pedunculated hemangioma on the right posterior scalp without bleeding or ulceration (Figure 1–5). No other lesions were discovered elsewhere on his body. He had mild stertor at rest without significant retractions or increased work of breathing; however, work of breathing increased during bottle feeding resulting in spillage of formula from his mouth.

Overview

Hemangiomas are common vascular tumors, occurring in up to 10% of children.[1] They grow rapidly during the first year of life and depending on the anatomic location can cause significant functional and cosmetic impairments. Hemangiomas of the neck should be managed based on the size and symptoms (eg, ulceration, bleeding) of the lesion. Rapidly growing tumors of the anterior neck can cause compression and result in airway and feeding difficulties in young infants; therefore, prompt

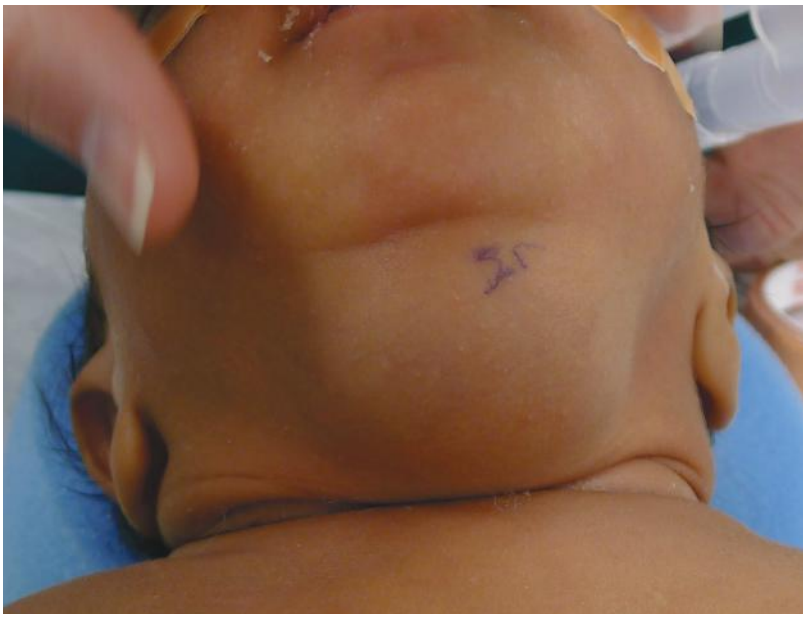

Figure 1–4. Midline anterior neck hemangioma.

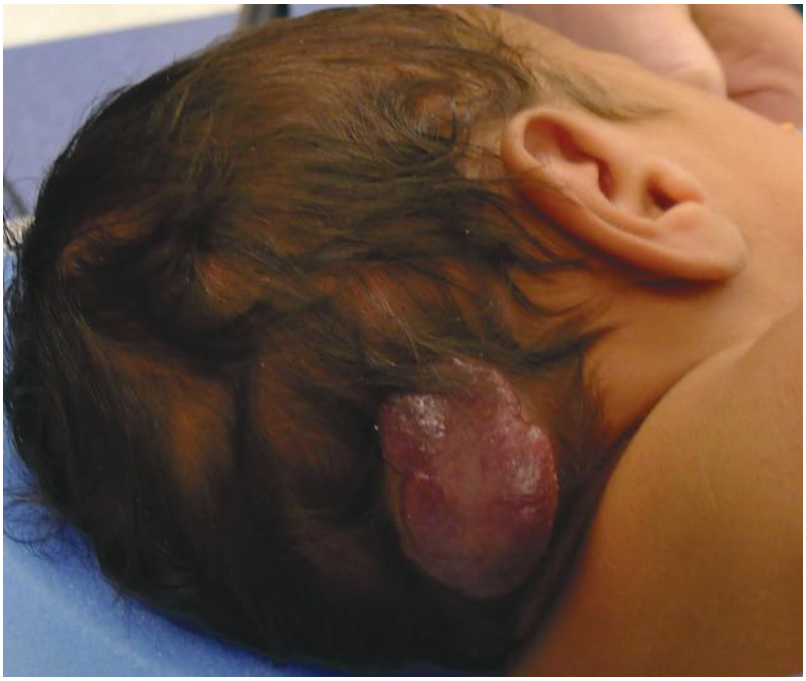

Figure 1–5. Occipital compound hemangioma.

diagnosis and appropriate medical or surgical treatment are critical. Small lesions may be monitored clinically or managed medically with propranolol therapy, steroid injection, or pulsed-dye laser therapy. Large lesions, especially those with a deep component more than 2 to 3 cm, are often amenable to complete surgical excision with little to no associated morbidity.

Differential Diagnosis

1. Infantile hemangioma
2. Lymphatic malformation
3. Mixed lymphatic-venous malformation
4. Benign tumor (eg, dermoid cyst or thyroglossal duct cyst)
5. Malignant tumor (eg, rhabdomyosarcoma or neuroblastoma)

This case illustrates the importance of considering infantile hemangioma in the differential diagnosis of a child presenting with a neck mass. The depth (with little to no skin involvement) of the lesion, the midline location, and cystic appearance of the mass made the diagnosis difficult based on physical examination alone. However, the absence of the mass at birth, its rapid progression, and the coexistent compound hemangioma on the scalp with a similar growth pattern, were all important clues in making the final diagnosis.

Diagnostic Workup

History

Information regarding the timing of onset and growth pattern of the neck mass, as well as associated symptoms such as fever, pain, bleeding, and overlying skin changes should be obtained from the caregiver. Birth history is important to note as well, as hemangiomas are more common in premature infants and infants from multiple gestation pregnancies.[1] The caregiver may describe snoring, stridor, or feeding difficulties depending on the size and location of the lesion. Any previous treatments or diagnostic tests should be considered.

Physical Examination

A complete head and neck examination should be carried out on any child presenting with a neck mass. A full body examination should be prompted if hemangioma is suspected, as patients will often present with multiple lesions. Careful inspection of any lesions should be performed, noting specifically size, depth, and skin involvement. Deep lesions will be completely covered with skin or mucosa and may or may not show subtle color change; compound and superficial lesions will have a red, blue, or purple color change to the epidermis. Auscultation of the chest can reveal any abnormal breath sounds that may suggest airway compression from the mass. When awake, flexible fiber-optic laryngoscopy (FFL) can be performed for further evaluation if there is suspicion of airway involvement.

Tests

Ultrasound is considered the initial imaging modality of choice in children presenting with a neck mass as it can usually be performed quickly and safely without the need for sedation.[2] In this case, important characteristics of the mass were seen on ultrasound (hypervascularity and calcifications)[3] that further supported the diagnosis of hemangioma and obviated the need for further imaging. The mass was limited to the anterior neck, and no other mass lesions were seen. Computed tomography (CT) or magnetic resonance imaging (MRI) could be used if further anatomic detail is needed, but it is not required.

Case Management

On initial presentation, the infant was already showing some signs of airway compression and feeding difficulty from the rapidly growing neck mass. The compound hemangioma of the posterior scalp had a fragile superficial component that was at increased risk for bleeding and ulceration due to its location and rapid growth. Therefore, surgical management was recommended.

The patient was taken to the operating room for excision of the anterior neck and occipital scalp hemangiomas. The scalp hemangioma was addressed first. A horizontal elliptical incision was designed to remove the hemangioma in its entirety. It was felt that there was enough laxity in the scalp tissue to perform complete excision and primary closure without excessive tension. After injecting the skin with 1% lidocaine with 1:100,000 epinephrine, a 15-blade scalpel was used to make the skin incision just lateral to the superficial component of the hemangioma. The superior portion of the ellipse was incised first, and a subcutaneous dissection was performed using the scalpel until the lateral extent of the lesion was identified. A plane was then developed between the galea and the tumor. The hemangioma was retracted inferiorly until the deep component was encountered. A nonstick bipolar cautery at a setting of 15 watts was used to dissect the deep component away from the underlying tissues. A portion of the deep component left behind was cauterized for hemostasis and to shrink the vascular tissue. Feeder vessels were encountered and easily ligated using the bipolar cautery. Excellent hemostasis was maintained throughout. The inferior aspect of the ellipse was then incised and a similar dissection was used with the bipolar cautery until the hemangioma was removed in its entirety. The wound, 5 cm in length, was irrigated and closed in layers using 4-0 PDS and 5-0 fast-absorbing gut sutures. A pressure dressing was placed at the end of the case.

Attention was then turned to the anterior neck hemangioma. A natural skin crease overlying the mass was injected with 1% lidocaine with 1:100,000 epinephrine and incised with a 15-blade scalpel. Subplatysmal flaps were elevated superiorly and inferiorly. The mass, with a gross appearance consistent with a hemangioma, was superficial to the mylohyoid muscles superiorly and the strap muscles inferiorly (Figure 1–6). A bloodless plane was developed deep to the mass, and nonstick bipolar cautery was again used to dissect along this plane, carefully dividing feeder vessels as they were encountered laterally (Figure 1–7). The mass extended inferiorly to the level of the cricothyroid membrane but with no involvement of the thyroid cartilage. The hemangioma was removed in its entirety (3 × 4 cm) with minimal blood loss. The wound was irrigated and closed in layers using 4-0 Vicryl and 5-0 monocryl sutures. A pressure dressing was used in favor of a surgical drain.

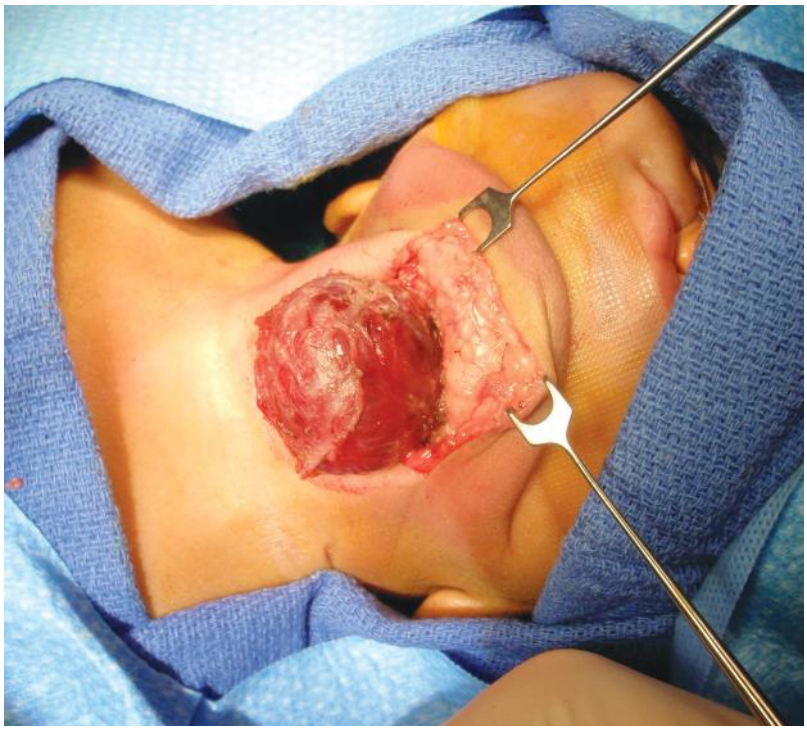

Figure 1–6. Subcutaneous midline neck mass with vascular appearance consistent with a hemangioma.

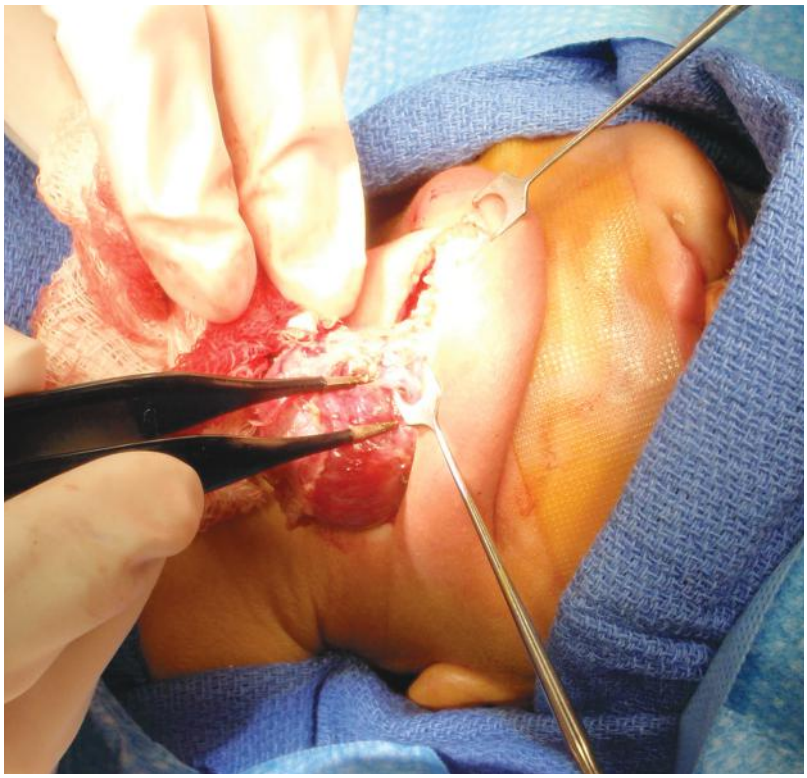

Figure 1–7. Bipolar cautery is critical to dissection of hemangiomas while maintaining excellent hemostasis.